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October 18- 20, 2018 Warsaw, Poland

Posters

Xin-Ye Jiang et al., J Pediatr Care 2018, Volume 4
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Mutational spectrum of phenylketonuria in Jiangsu province: Genotype–phenotype correlations and genotype-based predictions of BH4-responsiveness

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Phenylketonuria (PKU), an inherited congenital metabolic disease in newborns, is caused by variants in the phenylalanine hydroxylase (*PAH*) gene. A comprehensive analysis for phenylketonuria (PKU) variants can elucidate the mutational spectrum in patients from Jiangsu province, China. A total of 31 unrelated patients with PKU and their parents in Jiangsu, corresponding to 62 independent alleles, were investigated. Patients with BH4-cofactor deficiency were excluded. The patients were mainly from Suqian city in north of Jiangsu and Wuxi city in south of Jiangsu. These samples were collected from 2005-2012 via newborn screening program. All the patients studied were diagnosed as hyperphenylalaninemia at the Wuxi Maternal and Child Health Hospital, where phenylalanine levels on dried blood spots were quantified using the fluorescence test and tandem mass spectrometry. We systematically investigated 13 exons and their flanking introns of the phenylalanine hydroxylase (*PAH*) gene in 31 unrelated patients and their parents using the next-generation sequencing (NGS) technology. A total of 33 different variants were identified in 58 of 62 mutant *PAH* alleles. The variants c.721C>T, c.1068C>A, c.611A>G, c.1197A>T, c.728G>A, c.331C>T, c.442-1G>A were the prevalent variants with a relative frequency of 5% or more. One novel variant was identified in this study: c.699C>G. We studied genotype–phenotype correlation using the Guldberg AV system and revealed consistency rate of 38% (8/21) among the 21 predicted phenotypes. Genotype-based prediction of BH4-responsiveness was also evaluated and 14 patients (45.2%) were predicted to be BH4-responsive. The spectrum of *PAH* variants were presented in Jiangsu province and genotype-based prediction of BH4-responsiveness provides a useful tool in planning and management of future clinical trials using BH4.

Biography

Xin-Ye Jiang has completed her Bachelor's degree at Nanjing Medical University in 2003 and Master's degree in Pediatrics at Suzhou University in 2010. She is the Chief Physician of Child Hygiene in Wuxi Maternity and Child Care Hospital affiliated to Nanjing Medical University. She is currently serving as a Member of Child Health Professional Committee of Jiangsu Preventive Medicine Association and a Member of Child Health Group of Pediatrics Branch of Jiangsu Medical Association. She is mainly engaged in children's health clinical work and scientific research. For the last 5 years, she has headed and participated in six national and provincial research projects and has successively published 17 papers by the first author or the communication author, including three SCI papers and two Chinese papers.

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Notes:

Tereza Pomahačová, J Pediatr Care 2018, Volume 4
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Case report: Leopard syndrome in a newborn with severe hypertrophic cardiomyopathy

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We present rare form of RASopathy and myotonic dystrophy 1 with familial incidence. Our patient is a boy born at gestational age of 38 weeks by cesarean section to polymorbid tertigravida tertipara. After delivery there were signs of perinatal asfyxia, the prompt resuscitation was started, due to the absence of spontaneous breathing activity endotracheal intubation was performed. APGAR score was 2-4-7, an umbilical cord pH was 7,24. The neonate had clinical signs of fetal hydrops (ascites, pleural effusion, skin edema), the other clinical findings were gothic palate and equinovarus feet. Cardiac examination at 1st day of life revealed extreme hypertrophy of interventricular septum, decreased systolic and diastolic function. The heart function as well as pulmonary hypertension was improving according to regular cardiac examinations, ventilatory support was terminated after 18 days. 21st day of life an attack of ventricular tachycardia occured in the newborn, terminated spontaneously. The beta blocker therapy was initiated, with no recidive of arytmia. Based on the progressive ventricular hypertrophy the prognosis of the baby was uncertain. Leopard syndrome and myotonic dystrophy 1 were diagnosed prenatally. The patient was dismissed at age of 2 months in stabilized health condition, due to the severe cardiomyopathy he received DNR.

Biography

Tereza Pomahačová has completed her MD at the age of 25 years from Charles University – Faculty of Medicine in Pilsen. Since graduation in 2017 she has ben working at Department of Neonatology in Faculty hospital in Pilsen. Supported by a Grant from Ministry of Health of the Czech Republic – Conceptual Development of Research Organization Faculty Hospital in Pilsen – FNPI 9608.

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Ali Jangjou, J Pediatr Care 2018, Volume 4
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Reduction of the need for laparotomy considering clinical and laboratory findings in abdominal blunt trauma patients referred to Rajaei Hospital in year 2014 and 2015

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Background & Aim: Trauma is the most common cause of death in persons aged 1-44 years and the third most common cause of death regardless of age. Abdominal trauma is one of the most prevalent cases of trauma. Laparotomy is a therapeutic method which is applied for abdominal trauma. The aim of this study was prediction of the need for laparotomy considering clinical and laboratory findings in abdominal blunt trauma patients referred to Rajaei Hospital.

Materials & Methods: This retrospective study was done on all patients who had undergone emergency laparotomy at Shahid Rajaei Hospital at Shiraz for abdominal trauma during, the years 2014-2016. The data were collected through a questionnaire. The statistical analysis was performed using the SPSS 21. P value was significant if was less than 0.05.

Results: Of the 81 trauma patients who had undergone laparotomy, 66 cases (81.5%) was male and 15 (18.5%) was female. 95.1% of laparotomies were positive. Patients who had positive laparotomy had lower systolic blood pressure ($p=0.02$), Glasgow coma scale (GCS) ($p<0.001$) and hemoglobin ($p=0.03$) and higher respiratory rate ($p=0.04$). Spleen and liver were the common damage organs.

Conclusion: This study showed that blood pressure, GCS, hemoglobin and respiratory rate can predict the need for laparotomy.

Biography

Ali Jangjoo has completed his Postgraduation in Emergency Medicine at Shiraz University of Medical Science. He is the Director of Motahhari Clinic in Shiraz. He is an Assistant Professor at Shiraz University of Medical Sciences.

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Hyun-Duck Kim et al., J Pediatr Care 2018, Volume 4
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Association between periodontitis and early atherosclerotic vascular disease

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Background & Aim: We aimed to evaluate the association of periodontitis with the development of early atherosclerotic vascular disease in Korean adults.

Methods: In this cross-sectional study, a total of 1343 adults aged over 40 years were recruited from a community-based cohort of Yangpyeong County, Korea, during the period 2010e2014. Only dentate individuals were included in the study. Subclinical atherosclerosis (SA) was defined as carotid intima-media thickness (cIMT) of 0.754 mm, as assessed bilaterally by B-mode ultrasound. Peripheral arterial disease (PAD) was defined as ankle-brachial index (ABI) of 1.0, as measured by Doppler. History of periodontitis was assessed by measuring the radiographic alveolar bone loss (RABL) on a digital dental panorama and was classified into three groups: normal, moderate and severe periodontitis (two or more nonadjacent interproximal sites with RABL of 4 mm and 6 mm, respectively). The associations of periodontitis with SA and PAD were evaluated by multivariable logistic regression analysis and analysis of covariance, adjusted for age, sex, education level, tooth loss, smoking, drinking, exercise, obesity, triglycerides, HDL, LDL, hs-CRP, diabetes and hypertension. Stratified analyses were performed to identify specific risk groups.

Results: After controlling for confounders, severe periodontitis was associated with SA [adjusted odds ratio (AOR=1.55); 95% confidence interval (CI): 1.07e2.24] and PAD (AOR=2.03; 95% CI: 1.05e3.93). These associations were highlighted in never-smokers. For increasing severity of periodontitis, the adjusted mean cIMT increased (p=0.011) while that of ABI decreased (p=0.033).

Conclusion: Our data showed that periodontitis is a substantially important risk factor for atherosclerotic vascular disease among Korean adults.

Recent Publications

1. Lee J H, Shin Y J, Lee J H and Kim HD (2018) Association of tooth brushing and proximal cleaning with periodontal health among Korean adults: Results from Korea National Health and Nutrition Examination Survey in year 2010 and 2012. J Clin Periodontol. 45:322-335.
2. Lee J H, Shin M S, Kim E J, Ahn Y B and Kim HD (2017) The association of dietary vitamin C intake with periodontitis among Korean adults: Results from KNHANES . PLOS One. 12(5):e0177074.
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4. Ahn Y B, Shin M S, Han D H, Sukhbaatar M, Kim M S, Shin H S and Kim H D (2016) Periodontitis is associated with the risk of subclinical atherosclerosis and peripheral arterial disease in Korean adults. Atherosclerosis. 251:311-318.
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Biography

Hyun-Duck Kim has completed his PhD at Duke University, USA. He is the Director/Professor at Duke University, USA. He has over 200 publications that have been cited over 200 times, and his publication H-index is 20 and has been serving as an Editorial Board Member of reputed journals.

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The need for critical care education for emergency nurses

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Over recent years' emergency departments become more crowded this poses an emerging threat to patient safety and could have a significant impact on the critically ill. Emergency departments see older, sicker and more critical patients than ever before. A lack of available critical care beds, and the need of proper discharge planning results in critically ill patients boarded in the emergency department. They often occupy the much-needed trauma rooms especially when critically ill or intubated. If a critically ill patient ends up intubated in the emergency department's trauma room the primary nurse better be no junior. But, due to the well-known nursing shortage and a lack of a formal progressive orientation packages, junior nurses often end up taking care of critically ill and intubated patients. This might result in a life-threatening situation. During my time served as a nursing manager of a trauma center in the Middle East I recognized the need for critical care education on a daily basis and therefore was determined to improve critical care nursing skills within my department. As the need for critical care education for emergency nurses / transport nurses in the Middle East is high I decided to found CritBox, a startup that is on the way to generate what is much needed, Critical Care education for nurses. Let me take you on a journey that seemed impossible.

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Violent side of pediatric trauma-priorities in pediatric care and emergency medicine

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Significant number of the children brought to emergency department and injury is one of the leading cause for which they seek emergency services. Injury especially in children is the leading cause of the death and disability. Severely injured child need immediate attentions for transfer to well-equipped healthcare facility to diagnose and manage the injuries in children. Children and adults are anatomically, physiologically and emotionally different from each other. Early recognition and treatment of life-threatening airway obstruction, inadequate breathing, and intra-abdominal and intra-cranial hemorrhage significantly increases survival rate after major trauma. The initial assessment and management of the injured child follows the same ATLS® sequence as adults: primary survey and resuscitation, followed by secondary survey. Life-threatening conditions are dealt with as soon as they are identified. Necessary imaging studies are obtained early. Constant reassessment ensures that any deterioration in the child's condition is picked up immediately. The secondary survey identifies other injuries, such as intra-abdominal injuries and long-bone fractures, which can result in significant hemorrhage. The relief of pain is an important part of the treatment of an injured child. Focused abdominal sonography for trauma (FAST) has become a useful part of the initial trauma evaluation. Computed tomography (CT) remains the gold standard for diagnosing abdominal injuries. Although CT detects most abdominal injuries, pediatric victims of polytrauma have near-normal vital signs even in the presence of significant blood loss, and can deteriorate rapidly. These children should be monitored with extra vigilance during transport to the CT scanner, in the CT scanner, and in the emergency room. After penetrating trauma, high likelihood of requiring surgical intervention timely and accurate assessment in the emergency department (ED), with appropriate resuscitation and stabilization either until hemodynamic stability or until the patient is transferred to the operating room (OT) for definitive management. A coordinated and organized approach between the ED, pediatric intensivist, surgeons, radiology, blood bank, and the OT is necessary.

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Priorities in pediatric care and emergency medicine

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The healthcare model based on evidence that medicine science is a worth and well established modus operandi. This ongoing model works adequately to approach the physical body, the dense aspect of the human being. However, humans are beyond physical structures and diseases are also linked to aspects beyond physical structures as well. It is priority to enlarge medical science horizon and develop clear thinking about forces and dynamics of illness in instances further on its physical-chemical presentation. Integrative medicine, no longer complementary or alternative in concept and in practice, comes to guide integration of medical cognitive approach among physical body, vitality, soul and spirit. Anthroposophy as a spiritual science presents conception and practice to support anthroposophic medicine as a consistent model of integrative medicine. This workshop propounds to bring healthcare professionals an idea about integrative medicine care based on anthroposophic medicine in practice; not as complementary therapies, but as integration of physical body, vitality, soul and spirit based on cognitive methods and expecting cognitive results. The intention is to draw attention to pediatric scientific integrative medicine as a field of great need of development. The objective of this study is to broaden the healthcare assistance beyond physical-chemical pathological perspective. Bring attention to integrative medicine as priority in pediatric care and emergency medicine in clinical and scientific fronts.

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A rare case of linear and whorled nevoid hypermelanosis with global developmental delay, scoliosis and retinal degeneration with dermoscopic features

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Only 40 cases of the rare sporadic linear and whorled nevoid hypermelanosis (LWNH) are described in medical literature worldwide. It is characterized by hyperpigmented, reticulated, streaky and whorled patches along Blaschko's lines, without atrophy or preceding inflammation. It reflects an underlying mosaicism and is occasionally associated with systemic abnormalities. A five year-old female presented in our institution with multiple uniformly hyperpigmented patches, some linear over right upper extremity, both lower extremities and trunk with midline demarcation. Lesions were unchanged and asymptomatic since birth. Dermoscopy showed linear brown streaks with alignment along Blaschko's lines over right upper extremity, reticular over right thigh and brown structure less zones interrupted by dotted perifollicular hypopigmentation over right posterior trunk and anterior thigh. Histopathology revealed basal layer hyperpigmentation, sparse superficial lymphocytic dermal infiltrates, melanocytic hypermelanosis and flat-topped papillomatosis. Hematological and biochemical tests revealed no abnormalities. The patient has decompensated thoracic scoliosis with 2 cm divergence from plumb line. Developmental pediatrics referral revealed assessment of global developmental delay and examination under anesthesia showed peripheral retinal degeneration on both eyes. While this rare occurrence presents a challenging situation and is one reason why data is lacking, affected individuals should be evaluated because of disease associations, including a search for developmental and growth delays, skeletal abnormalities, and other systemic abnormalities. Despite few associations from previous case reports, no promise of chromosomal abnormalities can be found using next generation or whole exome sequencing. Together with the fact that treatments have been tried without much success, makes it a necessity for reporting.

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The double burden of malnutrition in refuge settlements

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A growing number of refugee camps in Uganda as a result of war in Southern Sudan are facing a double burden of malnutrition, that is, the persistence of under-nutrition, along with a rapid rise of over-nutrition and non-communicable diseases such as diabetes, hypertension and coronary heart disease. This double burden of malnutrition has resulted from various factors including: a marked transition in dietary patterns over recent years (e.g. shifts to energy dense diets high in saturated fat, sugar, and refined foods, and away from plant-based diets); inadequate access to healthy food choices; declining levels of physical activity; and inadequate access to health care services as a result of displacement and broader social determinants. In refuge settlements and host communities in Uganda, in addition to the high levels of under-nutrition, substantial levels of overweight/obesity have also been observed. At the national level, 35% of children of are stunted. The prevalence is even higher in host communities where 40% of children screened are stunted. Many low- and refuge settlements and host communities are undergoing a nutrition transition associated with rapid social and economic transitions. We explore the coexistence of over and under-nutrition at the neighborhood and household level, in a refuge settlement setting in Uganda.

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Analysis of susceptibility genes of children with severe cutaneous adverse drug reactions

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Severe drug eruption (severe cutaneous adverse reactions, SCAR) refers to a group of life-threatening severe cutaneous adverse reactions of drugs characterized by the skin and mucous membrane involvement which includes Steven-Johnson syndrome (SJS) and toxic epidermal necrosis necrolysis (TEN), drug rash with eosinophilia and systemic symptoms (DRESS) and acute generalized exanthematous pustulosis (AGEP). Severe drug eruption in children is a kind of severe drug eruption occurring within the age group of 14 years old. Because of the particularity of children, they are different from adults. The pathogenesis of severe drug eruption has not been fully elucidated, but pharmacogenetics studies have confirmed that some SCARs caused by specific drugs are associated with individual human leukocyte antigen (human leukocyte antigen, HLA) allele types. Associations of abacavir and HLA-B*57:01, carbamazepine and HLA-B*15:02, allopurinol and HLA-B*58:01, dapsone and HLA-B*13:01 were widely reported and the effectiveness have been confirmed. It is possible to detect the specific risk loci to prevent the occurrence of severe drug eruptions. However, the HLA allele has its own characteristics such as population heterogeneity (i.e.,the same SCARs is different in different ethnic groups related to the HLA allele), non-uniqueness (i.e., the same HLA allele may be associated with different drug reactions, and different types of drugs reactions induced by the same drug may be associated with different HLA alleles). we need to find more relevant HLA risk loci that can trigger drugs-related SCARs and look for other susceptibility genes.

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Urticaria in children: Is it food allergy or more?

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Introduction: A food allergy component in children with skin conditions is estimated depending on the prevalence of these allergies and possible local foods implication. The objective is to assess clinical features in food-allergic children, which can be complicated by confounding factors such as eosinophil hyperactivation, intestinal parasitosis, or diamine oxidase (DAO) deficit.

Method: A sample of 68 children with urticaria (average age 4.24 yrs) was selected from the allergology practice from 2014-2016. Cutaneous testing to food allergens was performed and complete blood count, total IgE titer, specific food IgE (sIgE), eosinophil cationic protein (ECP), anti-Toxocara IgG and DAO activity were determined.

Results: Eosinophilia was present in 17 patients (25%), 8 with raised IgE. Total IgE were high in 26 patients (38.2%). Among the 32 patients with normal eosinophil (Eo) count and total IgE, 18 also tested negative for specific food IgE, while only two of the eight patients with high Eo count and raised total IgE presented a negative FX5 test. Milk sIgE were positive in 25 patients, egg sIgE in 13, nuts sIgE in 10 and flour sIgE in 9. As confounders, ECP was raised in 7 patients, all with severe skin symptoms (including 5 with normal Eo count), anti-Toxocara IgG in 4 cases, and DAO activity borderline in 2 cases, low in other 2.

Conclusion: ECP is a key marker for assessing the eosinophil-driven inflammation in close correlation with skin lesion severity. Milk allergy is most prevalent in children, followed by egg, nuts and flour allergy.

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PYLORIC STENOSIS

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Pyloric stenosis, also called infantile hypertrophic pyloric stenosis, is a condition caused by an enlarged pylorus. The pylorus is a muscle that opens and closes to allow food to pass through the stomach into the intestine. When this muscle becomes enlarged, feedings are blocked from emptying out of the stomach. The retained feedings cause the infant to vomit. There is no known reason for enlargement of the pylorus. It is more common in boys than girls and usually affects children who are born at full term. It rarely occurs in premature infants. Although not thought to be hereditary, pyloric stenosis occurs more commonly in children of parents who had pyloric stenosis themselves as infants. Infants with pyloric stenosis typically begin vomiting during the first month of life, but onset of symptoms may be delayed. The main symptom of pyloric stenosis is vomiting undigested breast milk or formula soon after a feeding. Vomiting usually begins at four weeks of age but can happen as early as two weeks after birth. Once vomiting begins it becomes more frequent, and severe, and is often described as "forceful" or "projectile an examination of the abdomen may allow the doctor to feel the enlarged pyloric muscle (called an "olive"). If the pylorus cannot be felt, pyloric stenosis can be diagnosed by ultrasound study or by x-rays taken after the infant drinks a liquid called "contrast." Pyloric stenosis does not get better by itself and must be corrected with an operation. The operation is called a "pyloromyotomy before surgery, dehydration and hypochloremic alkalosis must be corrected, generally with an initial normal saline fluid bolus followed by infusions of half-normal saline containing 5% dextrose and potassium chloride when urine output is observed.

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Necrotizing enterocolitis in a preterm infant newborn and role of feeding - an update!

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It is a clinical case presentation of a male preterm infant newborn (+31 weeks) that was delivered in our hospital and transferred to NICU because of prematurity, VLBW and need of respiratory support. Baby shortly underwent necrotizing enterocolitis (NEC) on fifth day of life shortly after starting of expressed milk feeding. It was early detected by the use of near infrared abdominal spectroscopy (NIRS). Baby was deteriorated clinically in a couple of hours and undergone intestinal perforation with peritonitis. So, abdominal exploration surgery with intestinal resection and end to end anastomosis was done urgently. Baby improved gradually and early feedings was started and gradually increased up to full feedings with the use of human fortified milk (HMF), probiotics and prebiotics. The study stated the evidence-based feeding strategy guidelines for necrotizing enterocolitis (NEC) among very low birth weight infants and role of trophic feedings, probiotics, prebiotics and micronutrients in prophylaxis, prevention and management of NEC. Prematurity is the single greatest risk factor for NEC and avoidance of premature birth is the best way to prevent NEC; the role of feeding in the pathogenesis of NEC is uncertain, but it seems prudent to use breast milk (when available) and advance feedings slowly and cautiously; NEC is one of the leading causes of mortality, and the most common reason for emergent GI surgery in newborns; NEC remains a major unsolved medical challenge, for which no specific therapy exists, and its pathogenesis remains controversial; a better understanding of the pathophysiology will offer new and innovative therapeutic approaches, and future studies should be focused on the roles of the epithelial barrier, innate immunity, and microbiota in this disorder; and bioinformatics modeling is a new emerging strategy aimed at understanding the dynamics of various inflammatory markers and their application in early diagnosis and treatment.

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Pediatric disaster response

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This topic prepares emergency managers, emergency planners, and members of public emergency departments like EMS, Fire, Police, Public Health, and Hospitals in the field of disaster response and preparedness work to effectively, appropriately, and safely plan for and respond to a disaster incident involving children, addressing the specific needs of pediatric patients in the event of a community based-incident that include pediatric-triage, mass sheltering, reunification planning and pediatric decontamination considerations. Presentation topics: Emergency Management Introduction, Pediatric Response Introduction, Planning and Response, Needs Considerations, Pediatric Triage, Pediatric medical responses, Pediatric Decontamination, Mass Sheltering.

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Effects of DOCK8 deficiency on IL-10 producing regulatory B cells

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Dedicator of cytokinesis 8 (DOCK8) deficiencies are characterized by recurrent infections, increased serum IgE levels, eosinophilia, and a significantly high risk of allergic and autoimmune manifestations. DOCK8 is a regulating factor of actin cytoskeleton proteins involved in the development and differentiation of B cells. Regulatory B cells (Breg) are potent negative regulators of antigen-specific inflammation and T-cell-dependent autoimmune diseases mainly through producing inhibitory cytokine interleukin-10 (IL-10). The precise signaling mechanisms required for Breg functions remain unknown. We sought to elucidate the effects of DOCK8 deficiency on Breg function in patients and DOCK8KO mice. DOCK8 deficient patients (n=3) have decreased percentage of IL-10+CD19+regulatory B cells compared with healthy controls. In DOCK8KO mice, the percentage and number of IL-10+CD19+regulatory B cells were reduced compared with WT mice after induced by OVA. In DOCK8KO-WT bone marrow chimera mice, it showed the decreased number of Breg, but for DOCK8KO-μMT (B cell deficient mice) bone marrow chimera mice showed the normal number of Breg. Adoptive transfer of DOCK8-/-CD4+ naïve T cells to CD4KO mice exhibited decreased Breg percentage. Finally, In vitro and in vivo administration of recombinant IL-21could restores the percentage of Breg, it might be caused by LPS-driven, but not IL-21-driven, STAT3 phosphorylation was defective in DOCK8KO mice. In conclusion, DOCK8 deficiency causes Breg intrinsic defect, as a result of abnormalities of IL-21-producing CD4+ T cells in DOCK8 deficiency.

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Childhood leprosy in post elimination era; a real challenge to face

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Introduction: Leprosy is a chronic infectious, disabling social disease. The most vulnerable group is children since they reflect disease transmission and the efficiency of disease control.

Objectives: To study the epidemiological and clinical trends of childhood cases of leprosy in Nepal during 2010-2016

Methods: A retrospective study was undertaken analyzing childhood leprosy (≤15 years) from national leprosy registry.

Results: A total of 19206 new cases of leprosy were registered during this period, of which 1177 cases (6.12%) were of children. National prevalence is below the leprosy elimination cutoff but the trend is gradually increasing in recent years (from 0.77 to 0.82/10000). Similar is with the new cases among the children reflecting the active transmission. The mean age of the children was 10.5 years. The average slit skin smear positivity was only 27% patients of the tested. The MB among child comprised of 38% and PB comprised 62% whereas total new MB cases were significantly higher (52%). Neuritis occurred in 21% and grade II disability at the time of diagnosis was noted in 3.5% of total patients including children. Lepra reactions treated in referral centre were observed with ratio of 2.75:1 between Type 1 and Type 2. There were no cases of relapse among children

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Juvenile onset Psoriatic arthritis (jPsA) share similar clinical characteristics with adult onset Psoriatic Arthritis (PsA) but children report longer delay in diagnosis

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To assess clinical and demographic characteristics retrospectively between those patients that had jPsA onset and those that had Psoriatic Arthritis (PsA) disease onset as adults. A registry for Spondyloarhtropathies (SpAs) aiming to assess patients' disease progression over time (ethics approval REC: 07/H0701/74) was used as a source for the information required for analysis. A questionnaire was given to be filed in by the patients when attending the clinic. The timing was as close to the first attendance as possible (if it was not possible to be at the first attendance). The questionnaire had semi validated questions and its validation has been published before. A total of 277 patients with established PsA were asked when did the disease start. From the total of 277 there were responses enabling analysis from 220 patients (79.4%). From those 220 patients (mean age 51.14 sd 14.2 range 18-88) 13 patients (5.9%) reported age of onset below the age of 16 and classified as Juvenile onset group; 32 patients (14.5%) reported age of onset between 17 and 25 years and classified as adolescent group; 161 patients (73.1%) reported age of disease onset between 26 and 64 and classified as adult onset, while 14 patients (6.3%) reported disease onset above the age of 65. With the aim to further analyze the adult onset group we divided the group in 4 groups according to the decade of their age of disease onset. These were; 26 to 35 years old, 36 to 45, 46 to 55 and 56 to 64 years old. Patients with reported age of onset disease above the age of 65 were classified as geriatric group. From the PsA group data from the juvenile onset PsA (jPsA) (n-13) were then compared with the adult group with the age of onset 56 to 65 years (n-22) as the numbers of patients in the latter group and the gender distribution were similar to those of the jPsA group. In the total of 13 patients identified with age of disease onset of below 16 years of age, data on arthritis, enthesitis, dactylitis, axial, peripheral disease or both, HLAB27 status, disease duration, delay in diagnosis and the indices BASDAI BASFI , sleep disturbance, wellbeing over past week and treatment effect were collected . Same data collected from the adult group and comparison took place between the 2 groups. Statistical analysis was performed using the SPSS statistical program and the differences between groups were calculated using chi square t tests. The male to female distribution according to the age of onset showed that there are slightly more boys in the juvenile group (M:F = 7:6) and equal gender distribution in the 56 to 64 age group (M:F 11:11). When we looked and compared the juvenile onset PsA group with the middle age 56 to 65 adult onset group similar disease characteristics were found as per table. The treatment that the patients were in was different in that more juvenile onset patients were on biologics, and immunotherapy medication while more adults were on NSAIDs (12 patients in the adult onset group (representing 54.5%) vs 3 patients in the jPsA onset (23%) and had surgery at the time of the assessment (5 adults (22.7%) vs 1 in jPsA (7.6%) (p=0.06). However the effect of treatment was similar.

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